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Newborn with complete double penis and two separate scrotums: A case report

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ABSTRACT

INTRODUCTION: We present a newborn with double penis and double scrotum as a part of a caudal duplication syndrome (CDS) which is a condition includes duplication of the distal organs of the body. It is crucial to have knowledge about it to be able to be identified.
PRESENTATION: A male newborn presented with double penis, double scrotum double urethra, double colon, and double imperforate anus. After work up a low descending colostomy was done (4 stomas of duplicated colon) and started feeding with normal passage from colostomy.
DISCUSSION: The cause of CDS is unknown many theories have tried to explain that, but the most accepted theory is failure of monchorial twins to separate completely. CDS may be associated with other congenital anomalies as imperforate anus, renal anomalies, and omphalocele.
CONCLUSION: CDS is a very rare condition which needs multidisciplinary team to manage and needs staged repair. Most pediatricians and pediatric surgeons are unable to diagnose it, we add a case of CDS to the literature.

1. Introduction

Caudal duplication syndrome (CDS) is an extremely rare condition that includes duplications of the caudal part of the body including genitourinary and gastrointestinal systems along with the vertebral column. CDS is first described by Dominguez [1]. We present a neonate with CDS with a double high imperforate anus and double scrotum. This case report has been reported in line with the SCARE criteria [2].

2. Presentation

A male newborn aged 6 h weighted 2.6 kg with no history of prenatal exposure and could not be diagnosed prenatally, was admitted to the NICU (neonatal intensive care unit) in our institution. He was born of a non-consanguineous marriage. On physical examination, the patient has a double penis (diphalus) with double pinpoint urethral opening, through both the patient micturates normally, double scrotum containing one palpable testis for each Fig. 1. He has two separated natal clefts with no anal orifice. Initial diagnosis of anorectal malformation with CDS is established. The Invertogram showed a high variety of anorectal malformation, the US revealed a hydronephrotic left kidney, his echocardiography showed no abnormalities. Subsequently, he underwent a low descending colostomy that revealed a duplicated colon(4stomas) Fig. 2. The patient started oral feeding and tolerating and pass regularly from the colostomy (The two proximal ends). The Subsequent distal Loopgram (from the distal two openings showed duplicated colon down to the rectum with no fistulae with the urinary tracts Fig. 3. The MRI (Magnetic Resonance Imaging) showed left moderate hydronephrosis due to left congenital PUJ (pelvi-ureteric junction) stenosis and duplicated urinary bladder. Also, there is a tethered cord with no duplication of the vertebral column Fig. 4. The patient discharged 3 days postoperatively for follow up and subsequent planning for a staged repair.

3. Discussion

Caudal duplication syndrome was presented in the literature by Dominguez [1] Nevertheless, the exact etiology still obscured. However, its embryological etiology is claimed to be conjoined twins with incomplete fusion of some areas of the body giving duplication of these parts or monchorial twins with incomplete separation lastly, fibrous band dividing the caudal embryo into two [3,4]. An event in early embryological development is suggested by Alfadhel et al., relying on formation of the pathology by the derivatives of the three germ layers [5]

In our case, we found two natal clefts with an imperforate anus (IA) which is commonly seen in CDS. Variant forms of imperforate anus were mentioned in cases of CDS, two anal openings on

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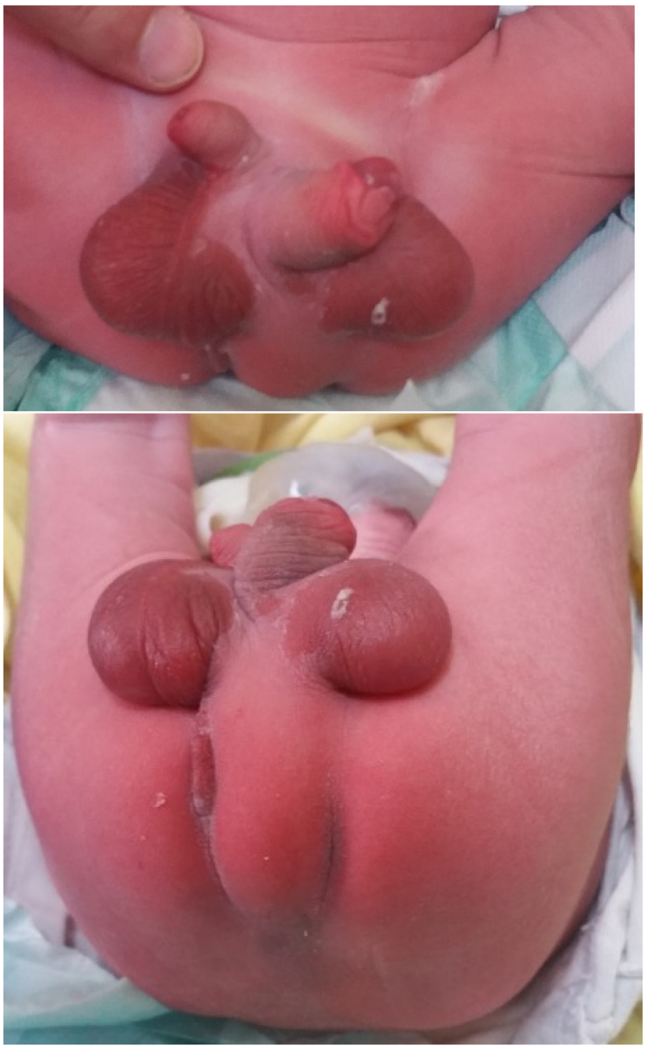

Fig. 1. Shows our patient with a double penis, a double scrotum, and two natal clefts with absent anal orifice.

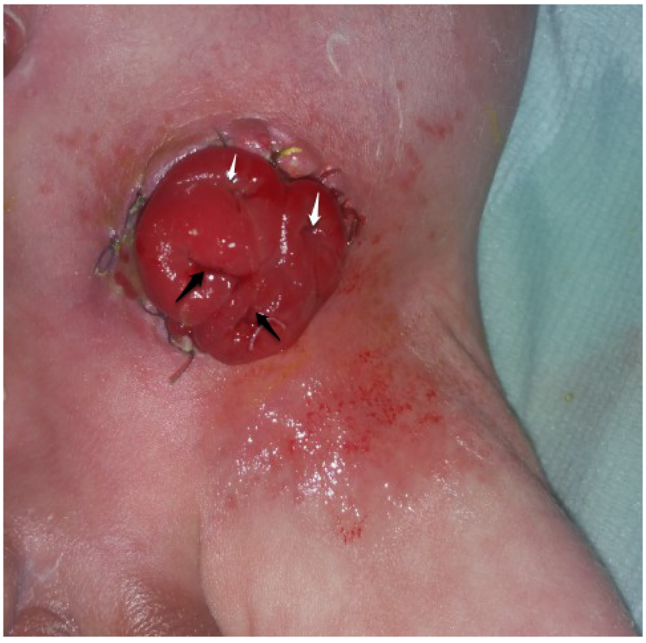

Fig. 2. Shows a low descending colostomy that demonstrates four stomas POD3: the two white arrows indicate the duplicated proximal colon and the two black arrows indicate the duplicated distal one.

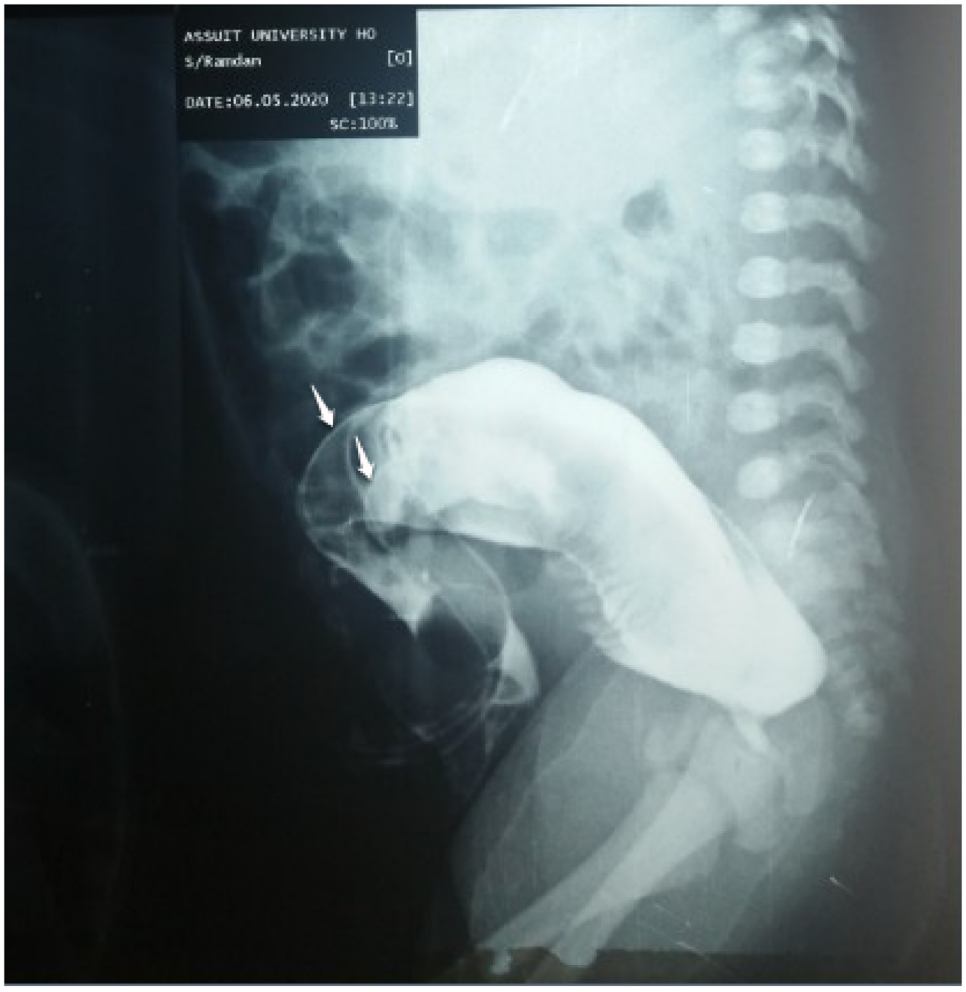

Fig. 3. Shows the distal loopogram detecting a duplicated distal colon: the two white arrows indicate the walls of the duplicated colon.

both sides, normal opening at one side, imperforate one at the other side and imperforate anus at both sides. Timing of occurrence of CDS as suggested by Bajpai et al. And Bannykh et al. [3] at 25th which falls in a time interval of cloacal membrane formation and its rupture (19th day and 9th weeks of gestation) [6] that can explain the increased frequency of IA with cases of CDS.

Double penis with two urethra and septated urinary bladder found in our case represents the caudal duplication of the genitourinary system in addition to a tethered cord and congenital left PUJ obstruction. Associations of these anomalies (caudal tubal duplication) in our case may favor the caudal twining theory as the mechanism behind CDS. Table 1 provides an overview of various cases of CDS reported in literature.

4. Conclusion

CDS is a rare condition that needs a multidisciplinary team for its management that includes staged repair of duplications. In our case, we are planning to start staged repair at the age of 16 months.

Written informed consent was obtained from the patient parents for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Table 1
cases with CDS in the literature.

Author(year)	Age	Gender	Anal region	External genitalia	Internal genitalia	Urinary system	GIS	Neurologic/vertebral system.
Suppiger(1876) [7]	NB	F	DIA and duplex Rectovulvar fistulas	DEG	DUt and DV	DUB and DU	DTC	Vertebral duplication (L3 and caudal parts)
Piccoli (1892) [8]	NB	F	Duplex anal orifices	Duplex vaginal orifices, one external genitalia	DUt and DV		DTC	
Volpe (1903) [9]	NB	M	DIA, right rectovesicular and left rectourethral fistulas	DEG		DUB and DU Horseshoe kidney	DC distal to cecum	
Bar et al. (1909) [10]	14 days	M	Blind anal dimple on each side of anal dimple	Bifid scrotum, one penis, two urethral orifices		DUB and DU	DC distal to cecum	Spina bifida, meningocele
Aitken (1912) [11]	NB	F	One normal anus and vulvar anus	Uterus unicornuate,			DTC + TI	
Lesbre (1927) [12]	?	F	Duplex anal orifices	DEG	DUt and DV	DUB and DU	DTC	Vertebral duplication (L3 and caudal parts)
Ombredanne (1936) [13]	2 years	F	Right normal anus, left rectovulvar fistula	DEG	DV	DU	DTC	
Aitken (1950) [14]	NB	F	DIA and duplex rectovaginal fistula	DEG	Bicornate uterus and DV	One urethra	DTC + TI	
Ravitch (1953) [15]	4.5 years	M				DUB and DU	DC distal to cecum	
Beach et al. (1969) [16]	?	F		DEG	DUt and DV	DUB and DU	DTC + TI	Meningomyelocele
Takaha et al. (1971) [17]	?	F	DIA	DEG	DUt and DV	DUB and DU	Duplication of rectum and appendix	Cleft sacrum, hemivertebra
Dutta et al. (1974) [18]	?	M		Duplication of penis, scrotum		DUB and DU	DC distal to midtransverse colon	Duplication of L5, split sacrum, hemivertebra
Veeraraghavan et al. (1983) [19]	?	M	DIA	Bifid scrotum		DUB and DU	DTC	Lateral deviation of sacrum, hemivertebra
Zamir et al. (1984) [20]	?	F		DEG	DUt and DV	DUB	DTC	Partial agenesis of the sacrum and coccyx, hemivertebra
Magalhaes et al. (1999) [21]	NB	F	DIA, left rectovulvar, right rectoperineal fistulas	DEG	DV	DUB and DU	DTC	Normal

Table 1 (*Continued*)

Author(year)	Age	Gender	Anal region	External genitalia	Internal genitalia	Urinary system	GIS	Neurologic/vertebral system.
Bannykh et al. (2001) [22]	NB	M	Right normal anal orifice, left imperforate anus with rectourethral fistula	DEG, left hypoplastic penis with hypospadias, right scrotal testis, left bilobed intraabdominal testis		DU	DTC + TI	Meningomyelocele, scoliosis, hypoplasia of left side of the lumbar spine, lumbar vertebra duplication, Sacral duplication
Kroes et al. (2001)-1 [23]	NB	F	Ectopic anus at right fold, A rudimentary, non-functional anus to the two vaginal openings	Normal urethral orifice, duplex vagina orifice	Duplication of cervix and DV (uterus unknown)	Pelvic right kidney, duplication of left ureter opening, urethra caudal to bladder neck, normal urethral opening	DTC	Complete duplication of the spine from L4 downwards, abnormalities in vertebral segmentation, abnormally shaped vertebrae and sacrum, myelocele
Kroes et al. (2001)-2 [23]	NB	F	DIA, rectoperineal and rectovaginal fistulas		DUt and DV?	DUB and DU, dilated pelvis of the right kidney	DTC + TI	Hemivertebra (T6, T10), abnormal curvature of sacrum
Bajpai et al. (2004) [3]	NB	M	Imperforate anus (high anorectal malformation)	Bifid scrotum, double phallus		DUB and DU, right VUR	DC from midtransverse colon to the rectum	Lumbosacral lipomeningomyelocele, lumbosacral spinal dysraphism, spina bifida
Vijayaraghavan et al. (2004) [24]	NB	F	Duplication of anus	?	DUt and DV	? DUB and DU	?	
Siebert et al. (2005) [25]	19-wk fetus	F	Imperforate anus	Vaginal atresia	DUt and DV	DUB and DU	DTC	Duplication of sacrum, coccygeal vertebrae, terminal spine
Jianhong et al. (2005) [26]		F	Right normal anal orifice, left low-level anal atresia	DEG	DV?	Bilateral complete duplication of kidney and ureter, DUB and DU	DTC	Normal
Liu et al. (2009) [27]	13 years	M	Two anuses beside perineal raphe, right anus was normal, left was closed as a small anal dimple	Diphallia (1 penile shaft, 2 glans)		DUB and DU, left hydroureteronephrosis	Duplication of stomach, duplication of transverse and sigmoid colon	Hemicorpus vertebral fusion of T11–T12, vertebral corpus subfissure of L5-S1

Table 1 (*Continued*)

Author(year)	Age	Gender	Anal region	External genitalia	Internal genitalia	Urinary system	GIS	Neurologic/vertebral system.
Taneja et al. (2009) [28]	NB	F	One anus, one urogenital sinus	DEG (urine output by the right orifice and feces by the left)	?	Pelvic left kidney, two urethral orifices, bladder diverticulum, left VUR	?	Complex malformations of the thoracic and lumbosacral spine, coccygeal vertebrae were absent, spinal cord duplication from T1, lipoma from L1
Alfadhel et al. (2009) [5]	NB	M	Right rectum opening to anus and left rectourethral fistula	Normal		DUB	DTC + TI	Left-sided lipoma at the tip of the conus medullaris
Bansal et al. (2011) [29]	2.5 years	F	Left normal anus, right anovestibular fistula	DEG	DUt and DV	DUB and DU, left anteriorly rotated pelvic kidney	DC with a single cecum	Scoliosis, lipomyelomeningo-cele, tethered cord and hydrosyrinx, Absence of right hemisacrum
Acer et al (2013) [4]	NB	F	DIA and duplex rectovulvar fistulas	DEG	DUt and DV	DU	Duplication of appendix, cecum, sigmoid colon and rectum	Vertebral dysplasias and bifid L5 and sacrum
Swaika et al. (2013)-1 [30]	NB	M	Duplex stenotic anal orifices	Two hemiphalus		DUB and DU	DC distal to caecum	Spinal lipoma
Al Alayet YF (2014) [31]	NB	M	DIA (right is low, and left is high with recto-bladder neck fistula)	Double perineum with DEG		DUB, Duplicated left pelvicalyceal system	DTC + TI	Duplication of lower 3 ribs, double pelvis, and tetrapagus.
Chaussy et al. (2015)-1 [32]	NB	F	Normal	DEG		DUB and DU	DTC + TI	hemivertebrae
Samuk et al. (2016) [33]	NB	M	Duplex anal orifices	Single penis with two Urethra and bifid scrotum		DUB and DU	DC with single caecum	splitting of the spinal vertebrae below the dorsolumbar spine, lipomyelomeningocele.
Our case	NB	M	DIA	DEG Double penis and double urethral orifices		DUB, DU, left congenital PUJ stenosis and ipsilateral hydronephrosis	DC to caecum	Tethered cord

NB: Newborn. M: Male. F: Female. GIS: Gastrointestinal system. DIA: Duplex imperforate anus. DEG: Duplication of external genitalia. DUt: Duplication of uterus. DV: Duplication of vagina. DUB: Duplication of urinary bladder. DU: Duplication of urethra. VUR: Vesicoureteral reflux. PUJ: pelvi-ureteric junction. DC: Duplication of colon. DTC: Duplication of total colon. DTC + TI: Duplication of total colon and terminal ileum.

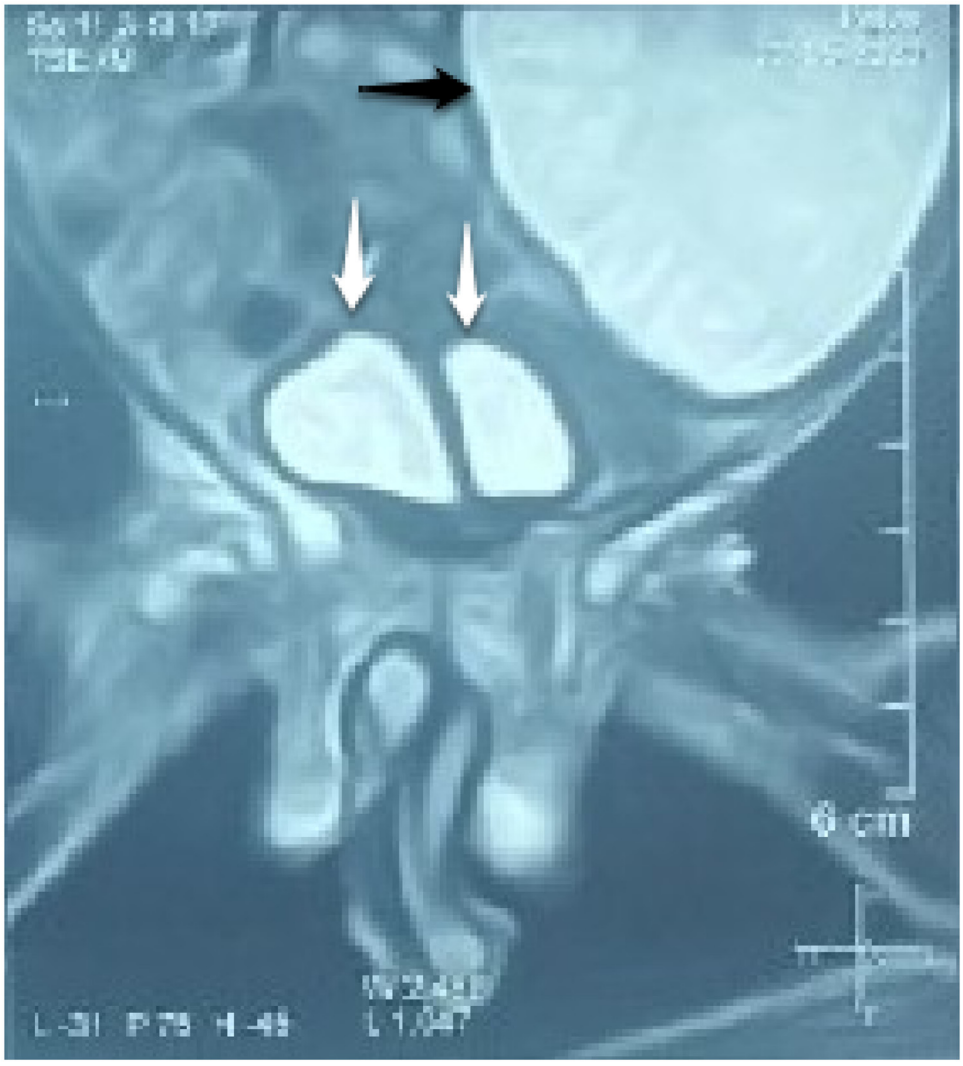

Fig. 4. Shows the MRI demonstrating a duplicated urinary bladder(the white arrows) and left hydronephrosis(the black arrow).

Declaration of Competing Interest

The authors report no declarations of interest.

Source of funding

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Ethical approval

The case report is exempt from ethical approval in my institution.

Consent

Written informed consent was obtained from the patient parents for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Ahmed Maher: main author of the paper and wrote the manuscript.
Hussein Ibrahim: literature review and revised the manuscript.
Mahmoud Mostafa: supervising and editing.
Tarek Abdelazeem Sabra: supervising and editing.

Registration of research studies

NA.

Guarantor

Ahmed Maher Ali Ahmed.

Provenance and peer review

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